Background: Inspiratory muscle strength in patients with neuromuscular disorders can be assessed using sniff inspiratory nasal pressure ($P_{n\text{sn}}$) and maximum inspiratory mouth pressure ($P_{\text{Imax}}$). However, the relative merits of $P_{n\text{sn}}$ against $P_{\text{Imax}}$ are not known in patients with severe neuromuscular disease. Objective: To investigate whether severity of disease modifies the relation between $P_{n\text{sn}}$ and $P_{\text{Imax}}$. Methods: Vital capacity (VC), $P_{n\text{sn}}$, and $P_{\text{Imax}}$ were measured in 258 patients with neuromuscular disorders. Results: Data were analysed from 241 patients, 17 being unable to perform $P_{\text{Imax}}$ or $P_{n\text{sn}}$ manoeuvres. The correlation between $P_{n\text{sn}}$ and $P_{\text{Imax}}$ was $r = 0.94$ ($p<0.0001$), with a mean (SD) difference between $P_{n\text{sn}}$ and $P_{\text{Imax}}$ of $4.8 (21.2)$ cm H$_2$O (the limits of agreement were 37.6 and $-47.2$ cm H$_2$O). VC (% predicted) was positively correlated with $P_{n\text{sn}}/P_{\text{Imax}}$ ($r = 0.86$; $p<0.0001$), with a lower $P_{n\text{sn}}/P_{\text{Imax}}$ value in patients with a VC $<40\%$ of predicted than in those with a VC $>40\%$ (0.80 (0.35) v 1.04 (0.41); $p<0.0001$). Conclusions: $P_{\text{Imax}}$ is greater than $P_{n\text{sn}}$ in patients with a severe restrictive ventilatory defect caused by neuromuscular disease. $P_{n\text{sn}}$ may not accurately reflect inspiratory muscle strength in such patients and it is thus advisable to use both tests.

**METHODS**

We studied 258 patients with neuromuscular disorders as part of routine clinical evaluation over a three year period at the Raymond Poincaré hospital. Seventeen patients were excluded: nine were unable to perform $P_{\text{Imax}}$ manoeuvres and eight were unable to perform $P_{n\text{sn}}$ manoeuvres.

All the tests were conducted in a single session, with patients in a seated position. $P_{n\text{sn}}$ and $P_{\text{Imax}}$ were both measured from functional residual capacity (FRC) in a standard manner, according to previously described methods ($P_{\text{Imax}}$ is an isometric manoeuvre, while $P_{n\text{sn}}$ is a quasi-isometric manoeuvre). $P_{n\text{sn}}$ was measured during 10 maximal sniffs, while $P_{\text{Imax}}$ was measured with a flanged mouthpiece with the manoeuvres repeated at least three times or until two identical readings were obtained. All signals were measured using a differential pressure transducer (Validyne, Northridge, CA, USA), amplified by a carrier amplifier (Validyne), and passed through an analogue-digital board to a computer running Acqknowledge software (Biopac System, Goleta, CA, USA). Patients received strong verbal encouragement with visual feedback, as previous studies have suggested. Spirometry and lung volumes were measured according to standard guidelines and reported as per cent predicted. Severity of disease was defined according to VC (% predicted), with a VC $<40\%$ being arbitrarily defined as severe restrictive lung disease. $P_{\text{Imax}}$ and $P_{n\text{sn}}$ are expressed as positive values although they are negative pressures with respect to atmosphere.

**STATISTICAL ANALYSIS**

All results are expressed as mean (SD). The correlation between $P_{n\text{sn}}$ and $P_{\text{Imax}}$ was assessed using linear regression analysis. Agreement between $P_{n\text{sn}}$ and $P_{\text{Imax}}$ was determined using a Bland and Altman plot. $P_{n\text{sn}}/P_{\text{Imax}}$ was used to evaluate the relations between $P_{n\text{sn}}$, $P_{\text{Imax}}$ and VC using linear regression analysis. The difference between $P_{n\text{sn}}/P_{\text{Imax}}$ in the patients with a VC $<40\%$ of predicted and patients with a VC $>40\%$ of predicted was assessed using an unpaired $t$ test.

**RESULTS**

Two hundred and forty one patients with neuromuscular disorders were eligible for analysis (table 1). Their mean (SD) age was 45.1 (16.4) years and 58% were male. No problems were encountered with air leaks during the $P_{\text{Imax}}$ manoeuvre using the flange mouthpiece. Mean VC (% predicted) was 51.9 (26.0)%, mean $P_{\text{Imax}}$ was 45.6 (28.2) cm H$_2$O, and mean $P_{n\text{sn}}$ was 41.5 (26.4) cm H$_2$O (35.2% of patients had a VC $<40\%$ of predicted and 37.8% were receiving non-invasive ventilation).

**Abbreviations:** FRC, functional residual capacity; NIV, non-invasive ventilation; $P_{\text{Imax}}$, inspiratory mouth pressure; $P_{n\text{sn}}$, sniff inspiratory nasal pressure; VC, vital capacity
As expected, there was a positive correlation between \( P_{sn} \) and \( P_{max} \) \((r = +0.94, p < 0.0001)\). However, the agreement between \( P_{sn} \) and \( P_{max} \) assessed using a Bland and Altman plot was relatively poor. Figure 1 shows a Bland and Altman plot with a mean difference between \( P_{sn} \) and \( P_{max} \) of -4.8 (21.2) cm H\(_2\)O and limits of agreement of 37.6 and -47.2 cm H\(_2\)O. Despite this, we observed a positive correlation between VC (% predicted) and \( P_{sn}/P_{max} \) (fig 2; \( r = +0.86; p < 0.00001 \)) which indicates that as VC falls the absolute value of \( P_{sn} \) declines more than \( P_{max} \)—that is, \( P_{sn} \) is greater than \( P_{max} \) in patients with VC less than 40% of predicted. Furthermore, the mean \( P_{sn}/P_{max} \) value was lower in patients with a VC less than 40% of predicted than in patients with a VC greater than 40% of predicted (0.86 (0.35) v 1.04 (0.41); \( p < 0.00001 \)). When expressed in absolute values (cm H\(_2\)O), \( P_{sn} \) was less than \( P_{max} \) in 147 of the 241 patients.

### DISCUSSION

As in previous studies of patients with neuromuscular disorders, we found a positive correlation between \( P_{sn} \) and \( P_{max} \), but with relatively poor agreement between these two tests. However, the present findings differ from previous studies in that the value of \( P_{max} \) was at least as great as, or even greater than \( P_{sn} \), particularly in patients with severe ventilatory restriction caused by neuromuscular disease. In fact, the present data are more consistent with, and extend, the observations of Landelli et al.\(^2\) and highlight the limitation of \( P_{sn} \) in this particular patient population.

Comparing our study with that of Stefanutti et al.,\(^1\) there are two major differences. The first is that overall \( P_{sn} \) is less than \( P_{max} \). The second is that advanced lung restriction from severe neuromuscular disease is associated with a lower value for \( P_{sn} \) than for \( P_{max} \). This disparity between the current study and Stefanutti’s could be the result of technical differences, as both \( P_{sn} \) and \( P_{max} \) manoeuvres in the earlier study were done without visual feedback. As sniffing is a more natural manoeuvre than \( P_{max} \),\(^11\) it is conceivable that without visual feedback \( P_{sn} \) would be greater than \( P_{max} \). In addition, in the Stefanutti study there were only three attempts at the unfamiliar \( P_{max} \) manoeuvre, but 10 sniff manoeuvres were done. In our clinical practice we get the patient to do at least three \( P_{max} \) manoeuvres or continue until two identical readings are obtained,\(^7\) and our greater \( P_{max} \) value may therefore be attributable to the learning effect. Furthermore, the current study differs from previous studies of patients with amyotrophic lateral sclerosis where facial muscle weakness and leaks around the mouthpiece were a major problem causing a reduction in \( P_{max} \).\(^11\)

The results of our study are supported by the findings of earlier studies in patients with chronic stable inspiratory muscle weakness\(^6\) and in patients with acute respiratory failure,\(^14\) showing that \( P_{sn} \) underestimates inspiratory muscle strength in patients with severe neuromuscular disease. Although we acknowledge that VC is an indicator of both respiratory muscle function and lung and chest wall compliance, it is more commonly used than \( P_{sn} \) or \( P_{max} \) in clinical practice to monitor sequential changes in respiratory impairment as neuromuscular disease progresses. Furthermore, in patients with advanced restrictive ventilatory defects secondary to neuromuscular disease there are marked reductions in VC with relatively small changes in maximum pressures, owing to the curvilinear relation between VC and maximum inspiratory pressures—that is, VC is probably a more sensitive marker of disease progression in advanced disease than in mild disease.\(^16\)

To explain the greater decline of \( P_{sn} \) than of \( P_{max} \) in patients with severe neuromuscular disease, we must

### Table 1: Patients with neuromuscular disorders eligible for analysis

<table>
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<tr>
<th>Diagnosis</th>
<th>N</th>
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<tr>
<td>Muscle</td>
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</tr>
<tr>
<td>Duchenne muscular dystrophy</td>
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<td>Becker’s muscular dystrophy</td>
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<tr>
<td>Nerve</td>
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<tr>
<td>Junction</td>
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<tr>
<td>Myasthenia gravis</td>
<td>32</td>
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</tbody>
</table>

![Figure 1](http://jnnp.bmj.com/)

**Figure 1** Bland-Altman plot: difference between sniff inspiratory nasal pressure (\( P_{sn} \)) and maximum static inspiratory pressures (\( P_{max} \)) plotted against the mean of these two variables \((n = 241)\). Filled symbols, patients with VC <40% of predicted; empty symbols, patients with VC >40% of predicted. VC, vital capacity.

![Figure 2](http://jnnp.bmj.com/)

**Figure 2** Relation between per cent predicted (% predicted) vital capacity (VC) and sniff inspiratory nasal pressure, normalised for maximum static inspiratory pressure \((P_{sn}/P_{max})\) \((n = 241\); \( r = 0.86; p < 0.0001\)). Horizontal dashed line indicates VC (% predicted) of 40% and the vertical line indicates unity between \( P_{sn} \) and \( P_{max} \) \((P_{sn}/P_{max} = 1)\).
consider the mechanism of sniffing compared with the maximal static inspiratory manoeuvre. Sniff is generated during a ballistic manoeuvre where the inspiratory muscles shorten to a greater extent and at a higher speed than during a $P_{\text{Imax}}$ manoeuvre, which is a more sustained isometric task. Thus as a result of both the force–velocity and force–length relations in striated muscle, $P_{\text{sn}}$ should be less than $P_{\text{Imax}}$—that is, pressure generation falls with a reduction in the operating length of the muscle and also with an increase in the velocity of muscle shortening. However, as sniffing is more natural than the maximum inspiratory static manoeuvre, it is believed that this accounts for the higher $P_{\text{sn}}$ than $P_{\text{Imax}}$ value in normal subjects. Also, the value for $P_{\text{sn}}$ is measured from the peak pressure, whereas $P_{\text{Imax}}$ is the average pressure sustained over one second, which includes an early peak pressure before falling off to a lower sustained pressure. Thus whichever of these two effects is greatest will influence the value of $P_{\text{sn}}$ more than $P_{\text{Imax}}$. Nevertheless, patients with severe neuromuscular disorders may not be able to perform a rapid sniff manoeuvre owing to significant muscle atrophy. Furthermore, because of the differences in the type of effort and the pattern of muscle activation in the two manoeuvres, the $P_{\text{sn}}$ value and $P_{\text{Imax}}$ value probably reflect different aspects of inspiratory muscle function. Thus a more sustained manoeuvre may achieve a greater pattern of inspiratory muscle activation in severely affected patients.

CONCLUSIONS

In patients with moderate to severe neuromuscular disease, overall $P_{\text{Imax}}$ yielded similar values to $P_{\text{sn}}$. Although there was a close correlation between $P_{\text{sn}}$ and $P_{\text{Imax}}$ there was a relatively poor agreement between the two variables, and the value for $P_{\text{Imax}}$ was higher than $P_{\text{sn}}$, particularly in patients with a severe restrictive ventilatory defect. $P_{\text{sn}}$ may overestimate the level of inspiratory muscle weakness, which challenges the view that this is the most appropriate test in this particular group of patients. We therefore suggest that $P_{\text{Imax}}$ and $P_{\text{sn}}$ are not interchangeable but are complementary tests and should be used in combination with VC for a complete sequential assessment of inspiratory muscle strength in patients with neuromuscular disease.

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